

Case report

**Cranio-facial fibro-osseous dysplasia. A case report and overview of the management of the condition**

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**Abstract**

Fibro-osseous dysplasia is a developmental, non familial, benign anomaly of bone development occurring in single or multiple bones, characterised by the replacement of normal bone by fibro-osseous tissue. We describe the case of a fourteen year old boy, which illustrates many of the difficulties confronting surgeons dealing with this uncommon condition. Complete surgical excision was required, before a definitive histological diagnosis could be established. We report the clinical, radiological and pathological features of this unusual case. We review the pathology of the condition and discuss the approaches to surgical management.

**Keywords:** Fibro-osseous dysplasia; Surgery; Pediatric

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## 1. Introduction.

While the term fibrous dysplasia of bone was introduced by Lichtenstein in 1938, the condition was first described by von Recklinghausen [32], with subsequent reports by McCune et al [1], and Albright et al [21] describing a syndrome associating fibro-osseous lesions with skin and endocrine abnormalities. Lichtenstein pointed out that the non-osseous lesions occurred in only three per cent of cases, and common to all these conditions was the abnormal replacement of bone with fibrous tissue. Lichtenstein and Jaffe's [19] report on 90 cases in 1942 led to universal acceptance of the term. However, controversy has since developed over the terminology and classification of fibrous dysplasia as it has emerged that the term encompasses a spectrum of pathological entities rather than a specific lesion [2]. Categorisation of any given fibro-osseous lesions requires correlation of the patients history and clinical, radiographic and operative findings, and histopathological features [33]. A useful working classification as proposed by Pecaro [24] is presented in Table 1.

Fibro-osseous dysplasia accounts for around 7% of all benign bone neoplasms [6], craniofacial involvement occurring in all of the severe forms and about 30% of the monostotic forms [11]. However, the distinction between monostotic and polyostotic forms can be very difficult [18] because of the intimate connection of the individual craniofacial bones and the fact that the disease does not appear to be limited by the suture lines. Fortunately, this distinction is not usually of great clinical significance. These conditions may not infrequently present to the otolaryngologist given the high incidence of craniofacial involvement.

There may be diverse presentations. Large areas of the skull may be involved [31], or an individual bone may be affected, typically the larger bones, such as the parietal bone [27], occipital bone [36] or most commonly the temporal bone [23]. Disease may be confined to a small craniofacial bone such as the sphenoid [10] or ethmoid bone alone [17]. Isolated involvement of the paranasal sinuses has also been described [16].

Table 1  
Fibro-osseous lesions of the head and neck (from [24]).

1.	Fibrous dysplasia
	Monostotic
	Polyostotic
2.	Fibro-osseous lesions arising from dental structures
	Periapical fibrous dysplasia
	Cemento-osseous dysplasia
	Ossifying Cementifying fibroma
3.	Fibro-osseous neoplasias
	Cementoblastoma Osteoblastoma (osteoid osteoma)
	Aggressive active ossifying fibroma

## 2. Case report

A fourteen year old boy with a seven year history of progressive nasal congestion was referred by his general practitioner for an otolaryngological opinion. The dominant symptom of obstruction was slightly worse on the right side and had proved resistant to various forms of medical therapy. There was no history of bleeding, pain, tenderness or nasal discharge. There was no previous history of trauma, surgery or foreign body insertion. His vision was normal and he did not suffer from headaches.

Outwardly the patient had a widened and flattened nasal bridge (Fig. 1a and b). Examination revealed a firm polypoidal mass almost completely occluding the right side of the nose with marked displacement of the anterior nasal septum to the left. Posterior rhinoscopy revealed that the mass also occluded the right posterior choana. Ophthalmic assessment confirmed normal conjugate eye movements with central uncorrected visual acuity in both eyes being J1. The peripheral visual fields and optic discs were also normal. The patient was anosmic, but neurological examination was otherwise unremarkable.

Routine biochemical indices, namely full blood count, E.S.R., urea and electrolytes and liver function tests including serum calcium, phosphate and alkaline phosphatase, were all considered to be within normal limits for his age, by our laboratory. Magnetic resonance imaging of the head revealed an enormous pear shaped tumour (Fig. 2) The superior narrow portion of the tumour occupied the mid-face and projected upwards as two domes into the anterior cranial fossa on each side. The lower extent of the tumour abutted the hard palate. There was displacement of each globe laterally. The lesion was of variable density, the upper portion appearing dense, while the lower portion appeared multilocular. This heterogenous pattern was confirmed on coronal CT scan which also demonstrated complete destruction of the cribriform plate (Fig. 3). Prior to operative intervention, bilateral carotid angiography was performed and revealed the tumour to be supplied by terminal branches of the ophthalmic artery, with an additional supply from the external carotid (Fig. 4). Repeated transnasal biopsies had failed to establish a diagnosis. It was decided to remove the tumour through a combined neurosurgical and rhinologic approach because the superior extent of the tumour could not be reached via a facial incision alone.

### 2.1. Surgery

A right lateral rhinotomy was performed to expose a tumour with a well-defined edge. The nasal septum was both eroded and displaced to the left. The lesion was of variable consistency and was removed piecemeal in it's lower two thirds. Surprisingly, both medial orbital walls were intact without bony erosion. Next, a right sided low frontal flap was performed and taken across the midline, to enter the frontal sinus. The frontal sinus was found to be filled with muco-purulent secretion, which was cleared and the lining curetted. The posterior frontal sinus wall was removed, the area irrigated with antibiotics and packed with Surgicel.

Having obtained good access, the dura over the frontal pole was opened bilaterally, and the frontal poles retracted to reveal intact dura covering firm tumour arising from below raising the dura as a dome, grooved in the midline, measuring two centimeters in height and three centimeters in width. This dome of Dura was incised and the underlying tumour removed. The Dura was then closed with Vicryl and covered with a free Xenoderm graft tamponaded with Surgicel between it and the brain. A bismuth iodoform paraffin paste pack was used to pack the nasal cavity.

The patient made an uncomplicated post operative recovery. After removing the nasal pack one week post operatively, the patient was discharged home on prophylactic anti-convulsant medication. At nine months follow up, he remains well.

Microscopic examination of the tissue fragments revealed rounded and curvilinear pieces of woven bone set in a moderately dense spindle cell stroma. Foci of calcification were present, which in areas formed rounded psammoma-like bodies (Fig. 5). Osteoblasts were inconspicuous. Areas of necrosis and inflammation were identified, in keeping with episodes of infection secondary to nasal obstruction. In some areas there was apparent infiltration of the nasal mucosa but no features of malignancy were identified.



Fig. 1(a).



Fig. 1. Pre-operative frontal (a) and profile (b) views demonstrating widening and flattening of the nasal bridge.

### 3. Discussion

#### 3.1. Pathology

Fibro-osseous dysplasia of the craniofacial bones represent part of a spectrum of benign lesions of uncertain aetiology. These are considered to be hamartomatous proliferation or localized failure of bone maturation rather than truly neoplastic lesions. There may be overlapping histological features. Common to these lesions are islands, or trabeculae, of new (woven) bone set in a connective tissue stroma of fibroblasts and collagen [24]. It is the degree of histological overlap in these lesions that can lead to difficulties in diagnosis and reinforces the need for correlation with the clinical and radiological findings.



Fig. 2. Midline sagittal T1 weighted MRI scans before (a) and after (b) intravenous Gadolinium injection. There is a mass within the ethmoid labyrinth protruding into the midline subfrontal region. Intermediate to low density before contrast, moderately enhancing after contrast injection. (c) Coronal proton density image.

The histopathological features that assisted in classifying this case as fibro-osseous dysplasia were that the trabeculae were composed predominantly of woven bone rather than mature lamellar bone, and the relative paucity of osteoblasts. These features along with the bland appearance of the stromal cells helped to exclude osteoma or a well-differentiated osteosarcoma from the differential diagnoses. The presence of the psammoma-like bodies coupled with the involvement of the cribriform plate raised the possibility of a meningioma of osteoblastic type but this was inconsistent with the radiological appearance.

Fibro-osseous lesions arising from dental structures could largely be eliminated on clinical grounds. The other main histological differentials—periapical fibrous dysplasia, cemento-osseous dysplasia and ossifying/cementifying fibromas—occur in an older age group.

Fibro-osseous dysplasia can present a complex histological picture which requires adequate, representative tissue with clinical and radiological information. The margins are often infiltrative and therefore biopsies from the periphery of a lesion may contain an admixture of reactive and lesional bone. Therefore foci of active osteoblasts (osteoblastic rimming) and lamellar bone do not preclude a diagnosis of fibro-osseous dysplasia in an otherwise clinically appropriate context. Areas of inflammation and haemorrhage may also complicate the appearances for similar reasons. As 'hybrid' lesions with a histological mixture of more than one fibro-osseous entity have been described [2], there is a clear case for pursuing a grand biopsy to effect a diagnosis as representative tissue cannot be guaranteed. The combined reference to clinical, radiological and pathological features is widely regarded as necessary to effect a diagnosis.

### 3.2. Radiology

Adequate imaging is invaluable in the management of this condition. As information toward staging, diagnosis and treatment planning is needed the bone detail from CT scanning [5] is particularly helpful. The presence of aneurysmal bone cysts is thought to be indicative of an aggressive lesion [35]. Serial imaging is mandatory and should be instituted the minute some type of skeletal dysplasia is suspected. It is particularly valuable in cases which are managed expectantly, to assess progression of disease. When operative treatment is considered arteriography is important to identify the vascularity of the lesion and its feeding vessels as major haemorrhage and postoperative intraosseous haematomas have been described [37]. Symptomatic multiple arterio-venous fistulae may also exist. [4]. Technetium<sup>99</sup> scanning is advocated by some as being useful in the localisation of individual lesions [14].

### 3.3. Treatment

Non-operative treatments would appear to be of limited value for these patients. The use of intra-venous pamidronate shows encouraging results but it is probably most useful in the severe systemic forms of the disease [20]. Steroids possibly, have a role in the management of the more painful cases. Radiotherapy has been

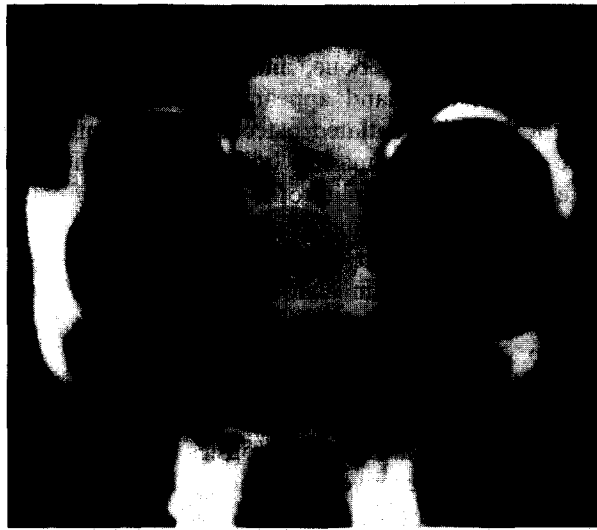


Fig. 3. Coronal CT scan. Calcium density mass occupying and expanding ethmoid labyrinth, encroaching on the maxillary sinuses and extending intracranially into the subfrontal region.

abandoned as a treatment modality in the condition, as the results were poor and the risk of malignant degeneration increased unacceptably [26]. Chemotherapy has proved to be ineffective in retarding disease progression [34].

It was previously considered prudent to wait for completion of skeletal growth as this might coincide with an arrest of further disease progression [13]. However, the growth rate of normal residual tissue and perhaps, more importantly, dysplastic tissue appears unaffected by early surgical intervention [30]. In some clinical situations, early surgery may prevent the onset of irreversible symptoms, such as loss of vision. The possibility of reactivation in later life exists however, especially in pregnancy [12]. Some authors have advocated that sequential radiographic monitoring may be adequate in many cases, with limited surgery for symptomatic complications [4]. Clearly these varied perspectives call for clinical judgement in any given case.

The management of the condition has undoubtedly become a subject of challenging complexity. The difficulties in establishing the surgical indications result from the benign nature of the lesion and the unpredictable natural history. These lesions can behave in a reactive, dysplastic or neoplastic manner [9]. There may be frequently sudden and irreversible symptoms that complicate the planning of surgical treatment beside the disease extent, especially with regard to involvement of the skull base [6].

A dilemma exists about the wisdom of early operative intervention. Resection may clear the disease but the impact of surgery on the maturing craniofacial skeleton must be considered [7]. An incomplete extirpation of the lesion may result in up



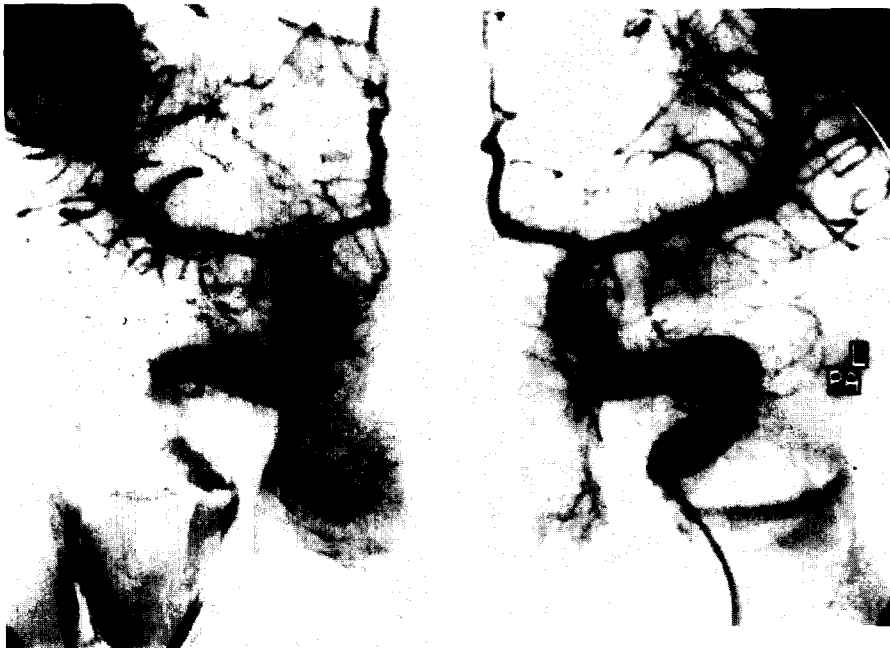


Fig. 4. Composite print of right and left anteroposterior internal carotid angiograms showing diffuse blush within mass lesion.

to a 25% local recurrence rate [15]. However, a more radical clearance may compound the difficulties of reconstruction. While an extended resection may help prevent the rare occurrence of malignant transformation in residual dysplastic bone, the extent to which this approach is justified remains uncertain.

We found the classification proposed by Chen and Noordhof [3] to be of practical value, in planning our surgical approach (Table 2). They suggest that surgery in patients in zone 3 should be avoided, until symptoms have actually appeared. They quote one exception to this, namely prophylactic deroofting of the optic canal, as once loss of vision has occurred, it is usually irreversible. Treatment of zone 2 lesions is optional as there is rarely any functional disability.

In the case presented, the patient's disease is classified as a zone 1 lesion. The primary aim of surgery was to correct the nasal obstruction, orbital hypertelorism and to prevent any further cosmetic deformity. A combined rhinologic-neurosurgical approach was chosen as it afforded the opportunity to achieve a complete exenteration, thus avoiding the possibility of later malignant degeneration. Cosmetically, it was felt that this approach would be acceptable to the teenage child (Fig. 6). Moore et al [22] state that in this group of patients (zone 1) radical excision of the pathological bone is seldom possible. It was most gratifying that a complete exenteration was achieved in this case.

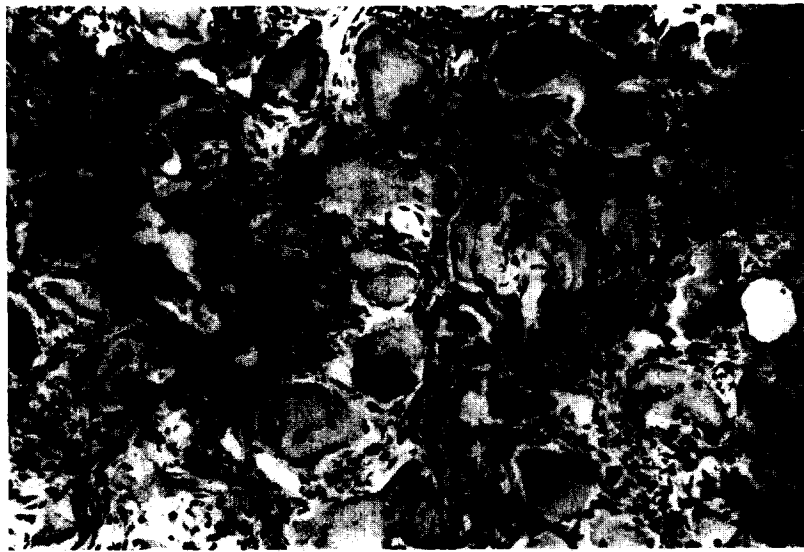


Fig. 5. Fibro-osseous dysplasia. Irregular islands of woven bone set in a moderately cellular fibrous stroma. A focus of calcification forming a psammoma-like body is seen. Haematoxylin and eosin  $\times 100$ .

Where the ideal aim of complete resection with minimal morbidity is not achievable, other surgical approaches may be utilised. Simple contouring of expanded bone back to normal dimensions, is effective in facial and skull bones despite a recurrence rate of between 50% and 75%, requiring revision surgery [25]. Lower recurrence rates have been ascribed to partial excision with normal autologous bone grafting. Donor site morbidity is avoided if acrylic implants [7] or blocks of resected, contoured, dysplastic bone [8] are used in reconstruction. This latter technique seemingly can be performed without the risk of apparent regrowth of diseased bone.

#### 4. Conclusion

Our case illustrates the principle that operative treatment must be tailored to the individual needs of the patient. The unusual histological variations in this case

Table 2  
Surgical classification (from [3]).

Zone 1	Frontal, orbital, nasal, ethmoid, zygoma, upper maxilla, (facial involvement amenable to surgery).
Zone 2	Parietal, part of occipital, temporal (lateral cranial base) (Hair covered cranium).
Zone 3	Central cranial base, petrous, mastoid, pterygoid, sphenoid (difficult or dangerous).
Zone 4	Maxillary alveolar bone, mandible (teeth bearing).



Fig. 6. Post-operative (nine weeks) frontal view.

highlights the heterogeneity of pathological presentation that may occur in these cases. It was satisfying that all the aims of surgery could be achieved in this case as this is rarely possible in fibro-osseous dysplasia.

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